CASE REPORT

ANDROLOGY

Familial appearance of congenital penile curvature – case history of two brothers

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Wojciech Zachalski Department of Urology Medical University of Gdańsk 17, Smoluchowskiego Street 80-214 Gdańsk, Poland phone +48 58 349 31 60 wojciech.zachalski@gmail. com The true prevalence of congenital penile curvature (CPC) is difficult to determine. Some study reports suggests that this problem may occur in as many as 10% of the male population [1]. However, a literature search of the Medline database revealed no reference concerning familial appearance of congenital penile curvature. For that reason we would like to present our case series.

Two brothers aged 25 and 26 respectively were admitted to the department of urology due to congenital penile curvature. Each patient was assessed by a history, physical examination, auto-photography of the erect penis, and a thorough sexual history. Concomitant anomalies of penile layers were absent in both cases. The Yachia [2] and Essed-Schroeder [3] corporoplasty technique were applied respectively. In follow-up both brothers reported straight erections.

A survey of the fetal penis at different stages of development shows some degree of curvature in a considerable number of embryos [4]. Penile curvature may thus be considered almost physiological in embryos between 35 and 45 mm in length. Thus, it has also been proposed that penile curvature is secondary to an arrest in normal penile development [5].

Therefore, some form of congenital local androgen deficiency may be responsible for inherited penile curvature.

Key Words: familial o congenital o penile curvature o corporoplasty

INTRODUCTION

The true prevalence of congenital penile curvature (CPC) is difficult to determine. Some studies suggests that this problem may occur in as many as 10% of the male population [1]. Severe penile curvature is associated with difficulty in vaginal intromission, discomfort to the patient or his partner during sexual intercourse, and psychological trauma.

Surgery remains the mainstay of treatment for patients with congenital penile curvature.

Various techniques have been developed for repairing penile curvature.

A literature search of the Medline database revealed no reference concerning the familial appearance of congenital penile curvature. For that reason we would like to present our case series.

MATERIAL AND METHODS

Two brothers aged 25 and 26 respectively were admitted to department of urology due to congenital penile curvature. Each patient was assessed by a history, physical examination, auto-photography of the penis, and a thorough sexual history. Concomitant anomalies of penile layers were absent in both cases. During outpatient preoperative evaluation patients were required to have three photographs (lateral, dorsal, and frontal) of the penis taken during erection (Figure 1).

Both patients were sexually active, but curvature resulted in difficulty or severe discomfort on vaginal intromission and partners' discomfort on vaginal intromission. On examination, the brothers were found to have se-

vere ventral (60°) and combined left ventro-lateral

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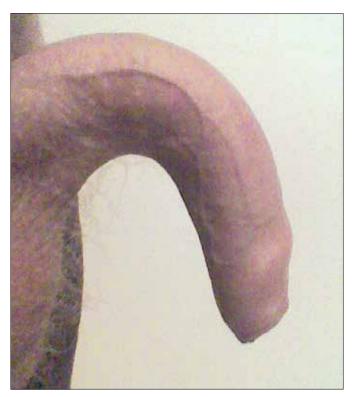


Figure 1. Preoperative autophotography picture from the side – ventral angulation.

(75° and 20°) curvature of the erect penis, respectively. Erectile function was normal.

Both were qualified for surgical correction.

In both cases, after degloving the penis with a tourniquet at the base, an artificial erection was created by injecting saline into the corpora cavernosa. (Figure 2)

A Yachia incisional corporoplasty [2] (Figure 3) was performed in the patient with ventral curvature (60°). Three pairs of longitudinal incisions of the tunica albuginea on the most convex side of the curvature in the dorsal part of the penis were made and then horizontally closed with continuous suture, Maxon 3.0.

In the second case (ventrolateral curvature), an Essed-Schroeder plicational corporoplasty [3] was introduced. Three pairs of reeving nonresorbable sutures were placed on the dorsal aspect of the tunica albuginea and one suture on the right hand side of the tunica albuginea to correct the curvature.

Circumcisions were performed to minimize the postoperative complication rate.

The kinship of the patients was independently verified by analysis of short tandem repeats (STRs) commonly used in forensic genetics. A total of 15 autosomal and 18 Y-chromosomal STR loci were amplified by multiplex polymerase chain reaction



Figure 2. Intraoperative picture taken after denudation – artifical erection.

and separated by capillary electrophoresis on a 3130 Genetic Analyzer (Applied Biosystems) (Tables 1, 2). Assuming prior probability of 50%, statistical analysis of the patients' genotypes revealed posterior probability of 99.999997% that they were full siblings, and thus confirmed their reported relationship.

RESULTS

Intraoperative check up revealed total straightening of the penis in both cases (Figure 4). The morbidity immediately after surgery was minimal. The patients' satisfaction with the cosmetic and

Table 1. Chromosomal sex assessment and autosomal STRs polymorphism

Locus	B.J	P.J
AMELX/Y	XY	XY
D7S820	9/10	9/10
CSF1PO	11/11	11/11
D3S1358	14/17	14/16
TH01	6/9.3	6/7
D13S317	8/11	8/12
D16S539	12/12	11/12
D2S1338	17/19	17/19
D19S433	13/15	13/15
VWA	15/17	14/17
TPOX	8/8	8/8
D18S51	15/16	14/15
D5S818	10/12	12/12
FGA	21/26	24/26
D8S1179	13/14	12/14
D21S11	29/29	29/29

Table 2. Y chromosome STRs polymorphism

Locus	B.J	P.J
DYS19	14	14
DYS388	12	12
DYS389I	12	12
DYS389II	29	29
DYS390	24	24
DYS391	10	10
DYS392	13	13
DYS393	13	13
DYS426	12	12
DYS437	15	15
DYS438	12	12
DYS439	12	12
DYS460	10	10
GATA H4.1	22	22
DYS385	11,14	11,14
YCAII	19,23	19,23

functional result was very good in long-term follow up (Figure 5). There was no evidence of persisting numbness of the glans nor erectile dysfunction after treatment.

CONCLUSIONS

The true prevalence of congenital curvature is difficult to determine. Some reports suggests that this problem may occur in up to 10% of the male population, however, clinically significant bending is much less frequent [1]. The most common type is ventral, but several different varieties exist.

The precise etiology of congenital curvature of the



Figure 4. Total straightening of the penis.

penis is unknown. During normal embryologic development, the penis goes through stages of ventral curvature and then straightens. Penile curvature may thus be considered almost physiological in embryos between 35 and 45 mm in length. Kaplan and Lamm have shown that 44% of embryos 100-150 mm long still maintain some penile curvature, which may persist after the 3rd month of pregnancy [5]. For this reason, penile curvature is frequent in severely premature babies and may show spontaneous remission with growth.

Embryologic development of the penis is regulated by testosterone that is converted by 5-alpha-reductase to dihydrotestosterone (DHT). DHT binds to androgen receptors in all tissue layers in the penis. Catuogno et al. postulate that fetal androgen deficiency or local deficiency of 5-alfa reductase may be the cause of different penile malformations.

By the external application of a suspension of DHT (0.15 mg/cm²) once daily for 2-3 months, they have



Figure 3. Intraoperative picture – Yachia incisional corporoplasty.



Figure 5. Picture taken six months after the procedure.

achieved almost complete straightening in five of 11 patients with penile deviations, but without simultaneous hypospadias and 30% improvement in two further patients [6]. Despite the fact that androgen deficiency may play a key role in the congenital penile malformations, we do not know what is the precise mechanism – environmental?, maternal?, fetal?, placental?

Histologically the deviation is associated with different abnormalities of the penile layers. Devine and Horton [7] proposed a classification system for congenital penile curvature based on that identifying three separate types of curvature. Type I is the most severe presentation. The structures normally surrounding the urethra (corpus spongiosum, Buck's fascia, dartos) are absent. Type II presents normal differentiation of the corpus spongiosum, but with fibrous formation of Buck's and Colle's fascia. In type III congenital curvature, the patient's urethra, corpus spongiosum, and Buck's fascia are all developed normally, but the dartos fascia forms the elastic

strip that causes the penis to bend. Kramer et al. [8] have proposed adding to the classification of Horton and Devine a fourth type, the curvature being due to asymmetry of the corpora cavernosa.

Microscopic study conducted by Darewicz et al. [9] revealed significant changes in the structure of the tunica albuginea such as chaotic alignment of the collagen fibers of different diameters, showing periodic widening and signs of disintegration and angulation.

In the two presented cases, the physical examinations showed penile curvature type IV according to extended Devine and Horton classification. Both patients were treated with standard procedures with good results. This is the first clinical report that attracts the attention of urologists to the familial form of penile deviation.

In our opinion further studies on CPC concerning epidemiology, genetics, histology, and embryology are needed. They may clarify if the CPC is a genetic disorder and what may be the best treatment.

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